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THE HEALTHY PKUER: WE ALL HAVE A ROLE TO PLAY

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This article looks at the role we all have to play in supporting PKU patients to reach and maintain their optimum low protein diet.

Specialist metabolic teams provide regular care and advice for PKU patients, but patients, their families; low protein food and protein substitute product manufacturers have a responsibility to support PKU patients in their lifelong low protein journey. Promoting and supporting a healthy low protein diet and lifestyle is everyone's responsibility.

PKUer *noun*: a person with Phenylketonuria (PKU). Often used in the low protein community by patients and parents. '*I'm a happy PKUer today!*'

Phenylketonuria (PKU) is an inherited metabolic disease, which causes disruption to the metabolism of the amino acid, phenylalanine (Phe). The lack of the enzyme, phenylalanine hydroxylase, which is necessary for the conversion of phenylalanine to tyrosine leads to hyperphenylalaninemia (raised Phe levels in the blood). When untreated, Phe accumulates in the blood with eventual excretion of phenylpyruvic acid in the urine.

If untreated, PKU leads to mental retardation and microcephaly.¹ Ongoing

uncontrolled Phe levels may result in further neurocognitive problems, such as reduced intelligence, poor attention span and impaired executive functioning.

In older children and adults, behavioural and psychosocial issues may present if the diet is poorly maintained.^{2,3} A wellmaintained low protein diet is of utmost importance in PKU females who are planning to be pregnant and during pregnancy, in order to prevent significant damage to the foetus. A low protein diet for life is highly recommended as treatment for PKU.

The low protein diet - the basics

A strict low protein diet is vital in maintaining low blood Phe levels and avoiding undesirable outcomes of the condition. The diet includes low protein food alternatives, such as bread, pasta, rice, baked goods; milk and meat substitutes; fruit and vegetables, plus amino acid or GMP based, phenylalanine free products (protein substitutes) in the form of liquids, powders and tablets. Rich protein food sources such as meat, fish, eggs, milk, cheese, nuts and seed; pulses and beans and soya, must be avoided.



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Exchange foods are included in the low protein diet and offer the small amount of natural protein that can be tolerated by PKU patients. Some fruits and vegetables must be limited, such as peas, sweetcorn, potatoes, bananas and dried fruits, but can be included as exchange foods. One exchange is equal to 1.0g protein (50mg Phe).

The amount of exchanges each patient can tolerate each day varies and can alter depending on age, growth status and type of PKU the patient has. Other exchange foods include breakfast cereals, yoghurt, crisps and snacks. For more details on exchange foods, please visit the NSPKU website: www.nspku.org/publications accessed.30/05/16>

Progress and development of the diet

Over the last 25 years, there have been huge developments in the kinds of low protein foods and amino acid or GMP based phenylalanine free products, which are available on prescription. Whilst this has vastly improved the choices for patients and the palatability of the diet, many PKU patients still struggle to find a balanced, healthy diet, which provides a satisfactory level of satiety and good quality nutrition.

The range of prescribed low protein foods is ever expanding, however. There are many products offering a sweet option, for example, low protein biscuits and cakes as snacks and low protein breakfast cereals are often quite sweet in comparison to standard healthier breakfast cereals, some of which can be included in the low protein diet as exchange foods. For example: Weetabix, 2.25g protein per biscuit = two exchanges or Porridge oats, 1.1g protein per 10g = one exchange*. Whilst these are healthy options, some patients may not have a great enough daily exchange allowance to include enough of them to fill up.

There have been calls from many metabolic dietitians for low protein food manufacturers to develop and produce low protein foods which offer healthier options, for example, low protein, low sugar, high fibre breakfast cereals, cereal bars or biscuits. In addition, a wider variety of low protein bread products, such as ready-made tortilla wraps, bagels, pittas, naans and chapattis, offer a high fibre content and provide further healthy variations to the existing range. A reduction in the sugar content and portion sizes of snack items, for example, cakes and biscuits, would further aid healthier low protein choices.

However, feedback from patients and parents regarding the choice of low protein foods currently available on prescription, calls for more convenience products, including low protein ready meals, low protein individually wrapped snacks and a wider choice of higher calorie foods such as donuts, crisps and desserts, products that reflect the current commercial food trends.

*Weetabix protein content available at www.tesco.com/groceries/product/details/ ?id=262146226 and Porridge oats protein content taken from Tesco Everyday Value Oats 1.0kg pack available at www.tesco.com/groceries/product/details/ ?id=259458142

The protein substitute products

There have been significant developments in the amino acid or GMP based phenylalanine free products that are available, where lower sugar and calorie versions are now available and palatability has been greatly improved. However, some PKU patients struggle to take their prescribed doses due to poor palatability, or they are deemed to be too high in calories and contribute to weight gain. Nevertheless, when compared with a recommended portion of protein rich food such as meat, fish, eggs, beans and pulses, these products are often lower in calories.

It is worthwhile discussing this with PKU patients, as they may not always be aware of the calorie value and how this fits in with their diet. Some patients find the volume of the products too filling, which can reduce the appetite for other foods.

PKU patients take their protein substitutes three to four times per day and it is recommended that they continue with their diet for life. Taste fatigue can be an issue; therefore, it is important to explore all of the product options with them. However,



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epresentative. *Lophlex LQ is the lowest volume ready-

hlex LQ is the lowest volume ready-to-drink protein substitute available for use.

there will be a patient who may benefit from the increased satiety of a product in order to avoid additional snacking.

The protein substitutes are an integral part of PKU management in that they aid control of blood Phe levels and provide the vital protein for growth, repair and immune functions, as the rich food sources of protein are strictly excluded. From an early age, these products should be seen as part of the diet rather than a medicine and continued education throughout the PKU management should stress the importance and benefits of taking the fully prescribed dose.

Overweight and obesity in PKU

Early studies looking at obesity in PKU patients concluded that overweight and obesity are common and the low protein diet, which is high in carbohydrates, may contribute to obesity in this patient group.⁴ However, more recent studies do not necessarily agree with this⁵ and there is some evidence to show that females with PKU are more at risk of becoming overweight or obese compared to males with PKU.⁴

Rocha et al⁶ studied the prevalence of overweight and obesity in PKU patients and it was found that there were similarities in the trends of overweight, obesity and body composition when PKU patients are compared with controls. Therefore, overweight and obesity trends in PKU are similar to that of the general population.

In light of these findings, it is acknowledged that managing overweight and obesity in the general population is challenging; however, PKU patients are well monitored, often by expert teams, who have a vital role in monitoring weight, nutritional status and general lifestyle choices, including physical activity.⁴

Although there is no evidence that documents best practice for managing overweight and obesity in PKU, the regular contact with healthcare professionals and metabolic company representatives should benefit PKU patients through promoting healthy dietary and lifestyle choices. Also any feeding problems and challenges should be discussed and treated to reduce the risks of overweight and obesity developing in this patient group.^{4,7}

The difference between low protein food and amino acid or GMP (glycomacropeptide) based phenylalanine free products:

Amino acid based - These products have been the mainstay for PKU management for many years. Made from synthetic amino acids, they offer all of the amino acids that the body needs, just without the phenylalanine. They have a distinctive aroma and taste. Many PKU patients can struggle with the bitter taste and this can influence compliance. Nevertheless, they are widely used. Lower calorie and sugar versions have been developed in line with current trends for a healthier lifestyle and weight management. Nutricia Metabolics, Vitaflo International and Promin (First Play Foods) all have amino-based products available for PKU management.

GMP based - These products have recently been approved for prescription in the UK. GMP (glycomacropeptide) is a whole protein, which is naturally occurring. It is created during the cheese-making process as one of the proteins within the whey. It is naturally Phe free. This protein is blended with large neutral amino acids (LNAAs) and arginine. The LNAAs reduce the absorption of the Phe in the GI tract and limit the passage of Phe in to the brain. They have a pleasant aroma and they are highly palatable. However, they may not be suitable for all PKU patients and may be too filling for some patients, who may struggle to consume their daily doses. Some PKU patients may benefit from the increased satiety achieved when consuming this type of product. Cambrooke Therapeutics provide a range of GMP products for PKU management.

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Questions relating to: The healthy PKUer: we all have a role to play Type your answers below and then print for your records or print and complete answers by hand.	
Q.1	What is Phenylketonuria (PKU)?
A	
Q.2	How is the condition treated?
A	
Q.3	Describe the low protein diet giving the foods that must be avoided
A	
Q.4	What is an exchange food and how is it measured? Give an example of an exchange food.
A	
Q.5	What role do the protein substitutes play in the low protein diet?
A	
Q.6	Explain how frequently PKU patients take their protein substitutes.
A	
Q.7	What do PKU patients struggle with when consuming their protein substitute?
A	
Q.8	Give the difference between amino acid and GMP (glycomacropeptide) products.
A	
Q.9	What does the current evidence say about the trend for overweight and obesity in PKU patients?
A	
Q.10	In PKU, explain which sex is more at risk of becoming overweight or obese and why.
A	
Please type additional notes here	

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